Radiology Case Reports 18 (2023) 1210–1216

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

Metaphyseal chondroblastoma in a pediatric patient☆,☆☆

Mauricio Paradeda Chiesa, MD[a], Breanna M. Thompson, BS[b], Natalia Maciel, MD[a], Dilia Diaz, MD[a], Pablo Stoppiello, MD[a], Leticia Gaiero, MD[a], Gottardo Bianchi, MD[a], Nicolas Casales, MD[a], Claudio Silveri, MD[a,], Ana C. Belzarena, MD, MPH, MBA[a]*

[a] *Unidad de Patología Oncológica Musculo Esquelética, Instituto Nacional de Ortopedia y Traumatología, Luis Alberto de Herrera 2226, Montevideo, Uruguay*

[b] *New York Institute of Technology, College of Osteopathic Medicine, 1855 Broadway, New York, NY 10023, USA*

ARTICLE INFO

Article history:
Received 10 December 2022
Accepted 27 December 2022

Keywords:
Chondroblastoma
Metaphysis
Pediatric patient

ABSTRACT

Chondroblastoma is a rare, benign neoplasm usually located in the epiphyses and apophyses of the long bones in the immature skeleton. Radiologically, these tumors have a classic appearance of a lytic lesion with chondroid matrix surrounded by a thin sclerotic rim. Here, we describe the case of a 5-year-old male who presented with a chondroblastoma unusually located exclusively in the metaphyseal region, which led to an elusive diagnosis. The presence of tumors outlying the traditional location or epidemiological spectrum, along with the potential for histopathological misdiagnosis, can pose a diagnostic and therapeutic challenge for the treating team.

Introduction

Chondroblastoma is a rare, benign neoplasm often located in the epiphyses and apophyses of long bones [1]. Representing approximately 1%-2% of all primary bone tumors, chondroblastoma occurs more commonly in children, adolescents, and young adults [2]. It characteristically affects males more than females at a ratio of 2:1 [3]. Chondroblastomas most frequently involve the proximal tibia followed by the distal femur, proximal femur and proximal humerus [4,5]. Patients commonly report gradual onset of bone pain, swelling and joint stiffness, and on physical examination decreased range of motion, muscular atrophy, and tenderness can be encountered [6]. Radiographically, chondroblastomas appear as an osteolytic lesion with a thin sclerotic rim, often involving the medullary cavity rather than the cortex [7]. Histologically, there is an increase in chondroblasts, the

☆ Funding: No funding was received for the purpose of this manuscript.

☆☆ Competing Interests: Each author certifies that neither he or she, nor any member of his or her immediate family, has funding or commercial association (consultancies, stock ownership, equity interest, patent/licensing arrangements, etc.) that might pose a conflict of interest in connection with the submitted article.

* Corresponding author.
E-mail address: claudio.silveri@gmail.com (C. Silveri).

https://doi.org/10.1016/j.radcr.2022.12.065

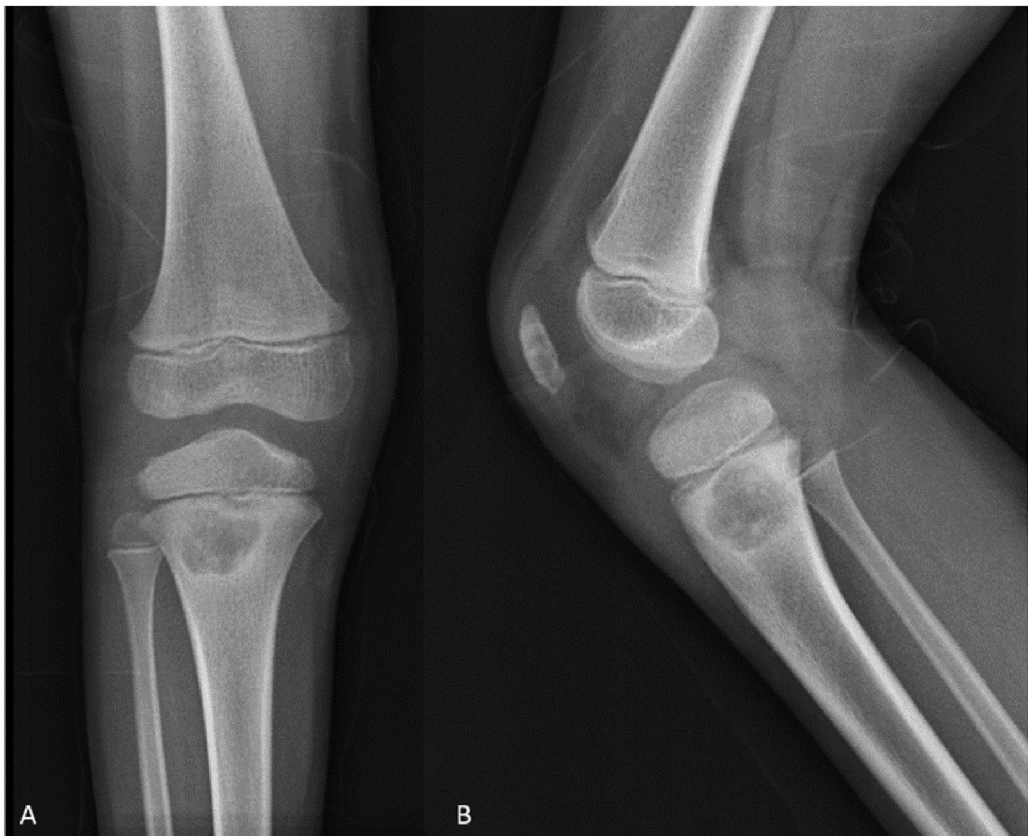

Fig. 1 – Right knee AP (A) and lateral (B) radiograph demonstrating a lytic eccentric lesion of the proximal tibia metaphysis surrounded by a thin sclerotic rim. The lesion does not invade the physis.

immature precursors to chondrocytes, and giant cells [3]. Chondroblastomas were found to express an H3F3B K36M point mutation which can aid in the correct identification of the tumor [8]. Intralesional curettage with adjuvants, such as phenol or hydrogen peroxide, is the gold standard for treatment [1]. Here we describe the case of a 5-year-old male who presented with a chondroblastoma unusually located exclusively in the metaphyseal region, which led to an elusive diagnosis.

Case report

A 5-year-old male without any relevant prior medical history, presented to the orthopedic service for consultation due to a chief complaint of right knee pain for the past 6 months. The pain had been intermittent and occurred without any trauma history. There was no associated fever or weight loss. The patient presented multiple times to the local emergency department (ED); however, no definitive diagnosis was made, and the patient was discharged on mild pain medication. During the last ED visit, a radiograph was obtained which demonstrated a proximal tibia metaphyseal lesion which prompted the orthopedic referral. The lesion was reported as having clear margins, with a mixed (lytic/blastic) matrix pattern, no periosteal reaction and no physeal involvement (Fig. 1). On physical examination during the orthopedic consultation the patient presented with no knee swelling, no skin changes, no local warmth or erythema, and positive tenderness to palpation of the proximal right tibia (Fig. 2). A decision was then made to request advanced imaging studies. On a CT scan without contrast the lesion presented with a cystic appearance and sclerotic margins; again, no physeal involvement or periosteal reaction was noted (Fig. 3). An MRI with and without contrast was also obtained demonstrating a multilocular cystic lesion with well-defined margins, fluid-fluid levels and contrast enhancement (Fig. 4). Given the patient's age, imaging characteristics and metaphyseal bone location the differential diagnoses of aneurysmal bone cyst and simple bone cyst were proposed. The patient was then taken to the operating room for a fluoroscopic guided percutaneous biopsy (Fig. 5). On histopathological analysis medium-size cells were observed with eosinophilic cytoplasm and chondroid matrix with calcifications and fibrosis. Occasional multinucleated cells as well as areas of pericellular calcification in a "chicken-wire" pattern were also present concluding the diagnosis of chondroblastoma of the proximal tibia metaphysis (Fig. 6). The patient then underwent a surgical procedure for curettage of the lesion with high burr as adjuvant, followed by filling of the area with bone allograft. At a 6-month follow-up the patient was pain-free, with full range of motion and radiographic healing of the lesion (Fig. 7).

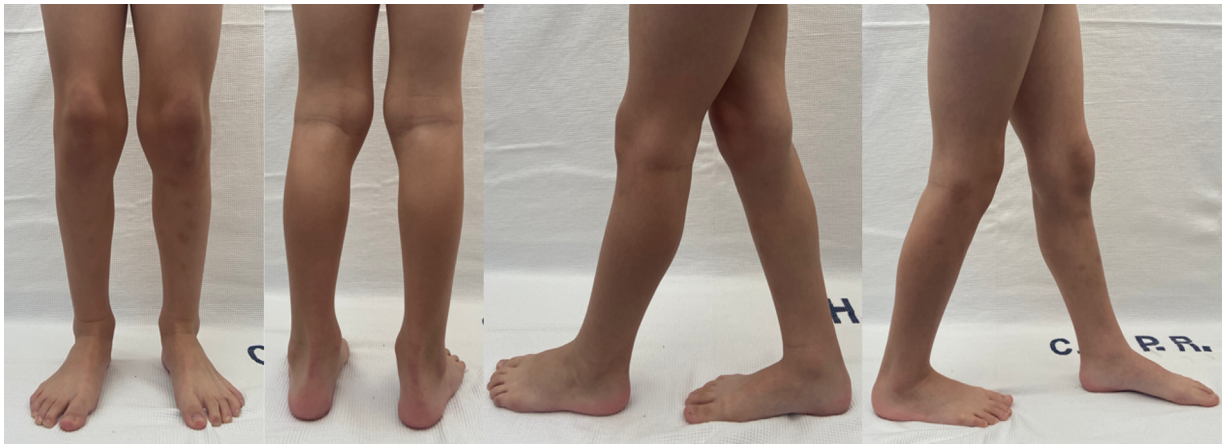

Fig. 2 – Clinical images of the patient on initial presentation at the orthopedic service demonstrating no inflammatory signs of the right knee.

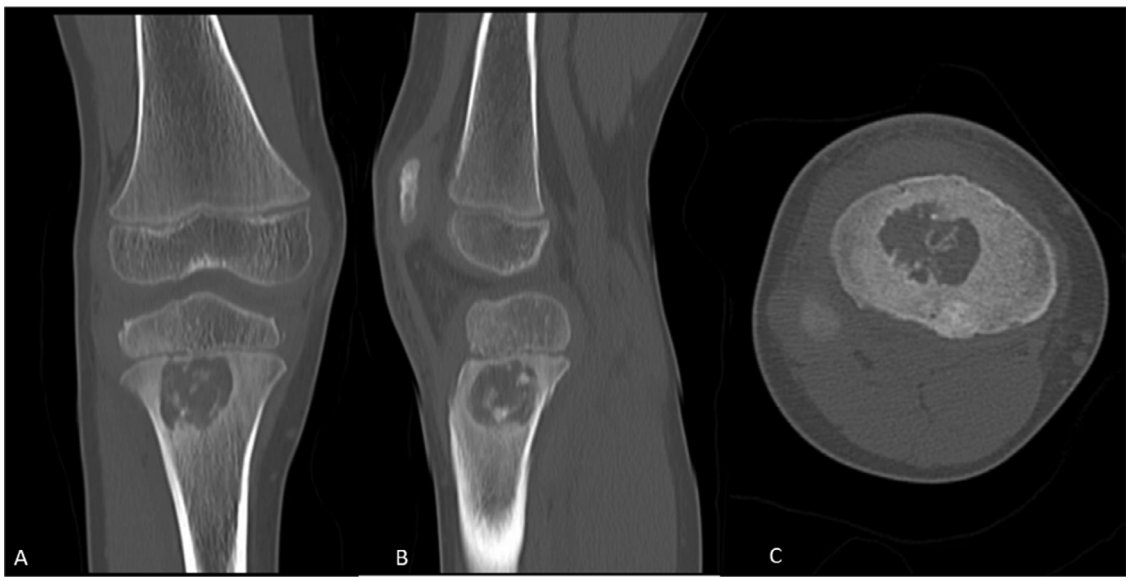

Fig. 3 – Right knee CT scan without contrast coronal (A), sagittal (B), and axial (C) views demonstrating a well-demarcated lytic lesion of the proximal tibia metaphysis with a chondroid matrix. No physeal involvement is noted.

Discussion

Chondroblastoma was initially described by Kolodny in 1927 and was followed by Codman who described the tumor in a proximal humerus location, which was since then designated as a "Codman's tumor" [9,10]. The lesion's main features are its location within a secondary ossification center in an immature skeleton and the presence of chondroblast surrounded by chondroid matrix with calcifications in a "chicken-wire" shape noted on histopathological analysis [1]. Even though most chondroblastomas are found in the epiphysis, 50% are capable of extending beyond the growth plate and into the metaphysis [5,11]. Less than 2% of chondroblastomas are located completely outside of the epiphysis, either on the diaphysis or the metaphysis of a long bone, with scarce cases reported in the literature, such as the rare case of our patient [12].

On imaging studies, chondroblastomas have a classic appearance which can easily suggest the diagnosis when present on a young patient and in the usual epiphyseal region [11]. The lesion is almost always in an eccentric intramedullary location, even though literature reports of extremely rare tumors in the intracortical setting exist [13,14]. On radiographic images, the tumor usually presents as a lytic lesion with well-demarcated margins, a thin sclerotic rim and chondroid matrix [15]. The area affected may be expanded, and in more rare circumstances, a cortical breakthrough with soft tissue extension may occur [16,17]. CT scans can reveal the lesion in more detail, demonstrating cortical thinning as well as intramedullary calcifications [18]. On MRI, these tumors are usually hypointense in T1-weighted images and

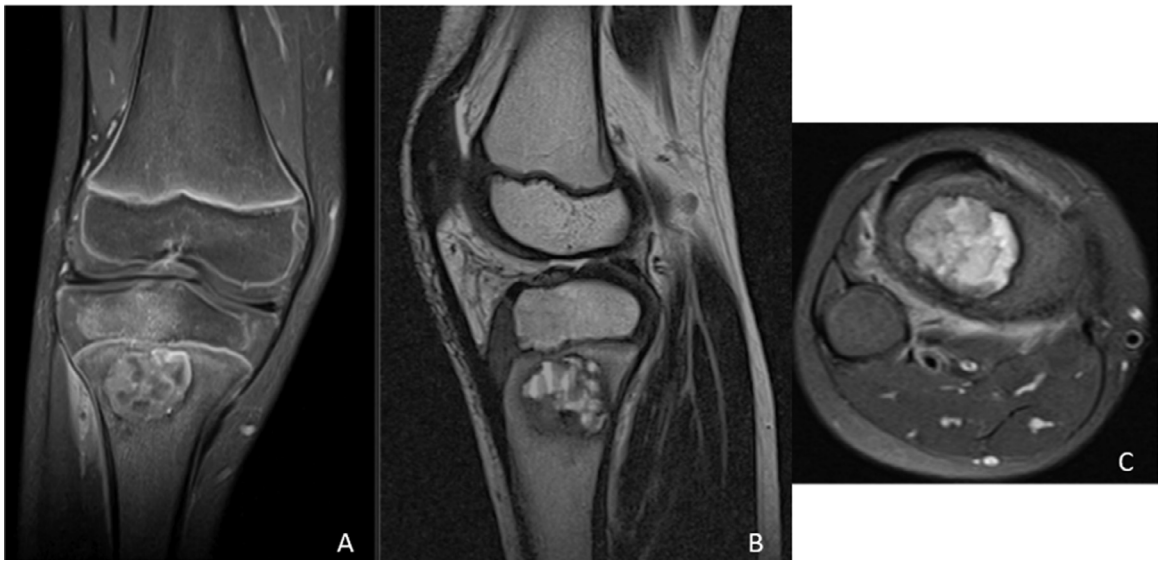

Fig. 4 – Right knee MRI with Gadolinium contrast coronal STIR (A), sagittal T1 (B) and axial T1 fat-suppressed with contrast (C) views showing a proximal metaphysis lesion with surrounding edema, fluid-fluid levels and heterogeneous enhancement.

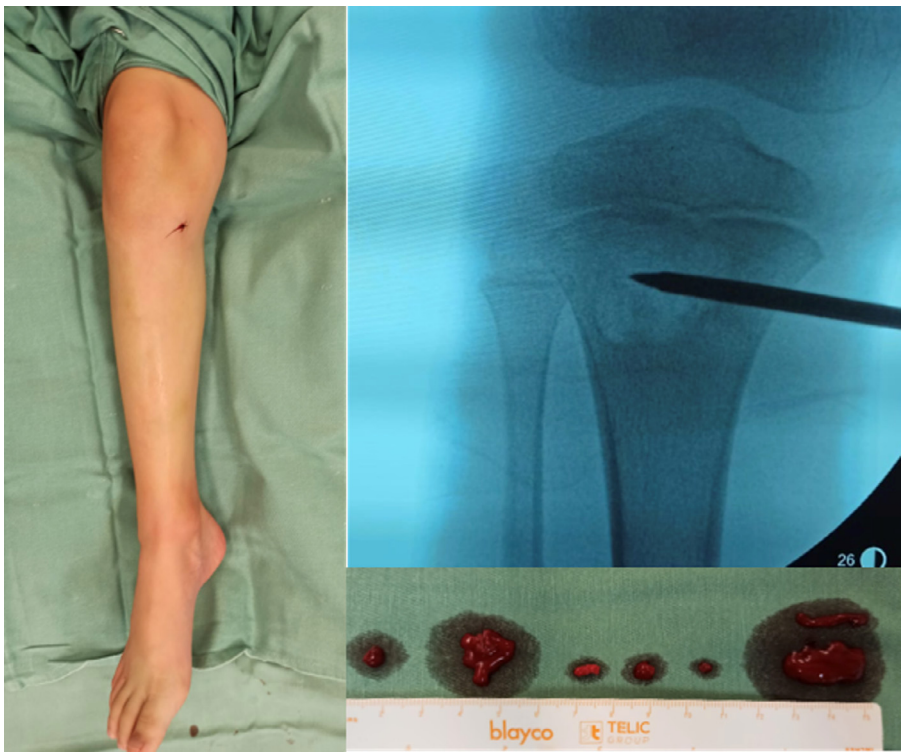

Fig. 5 – Images from the percutaneous image-guided biopsy and the gross sample obtained.

have a variable T2-weighted sequences appearance. Periosteal reaction as well as surrounding edema may be present. In Gadolinium-contrasted sequences peripheral, lobular and septal enhancement are noted [19].

Despite its oftentimes classical appearance, diagnosis must be confirmed by tissue sampling and histopathological analysis, especially in cases outlying the usual epidemiological spectrum or anatomic location. Furthermore, depending on the age of the patient, imaging features and bone region, several differential diagnoses can be proposed such as aneurysmal bone cyst, giant cell tumor of bone, chondroblastoma-like osteosarcoma, clear cell chondrosarcoma, and eosinophilic granuloma, among others [11]. On histopathological analysis, the lesion is characterized by

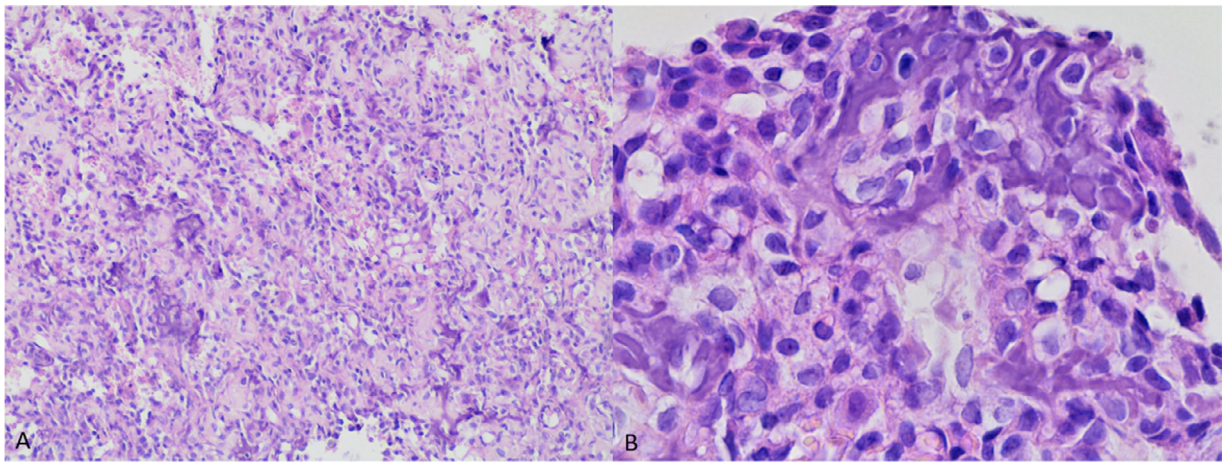

Fig. 6 – Microscopic image H&E x100 (A) and x400 (B) demonstrating a mesenchymal medium-size cell proliferation with mildly eosinophilic cytoplasm. Occasional giant cells and scarce chondroid matrix are observed. Fibrosis and calcifications in a "chicken-wire" pattern are also present.

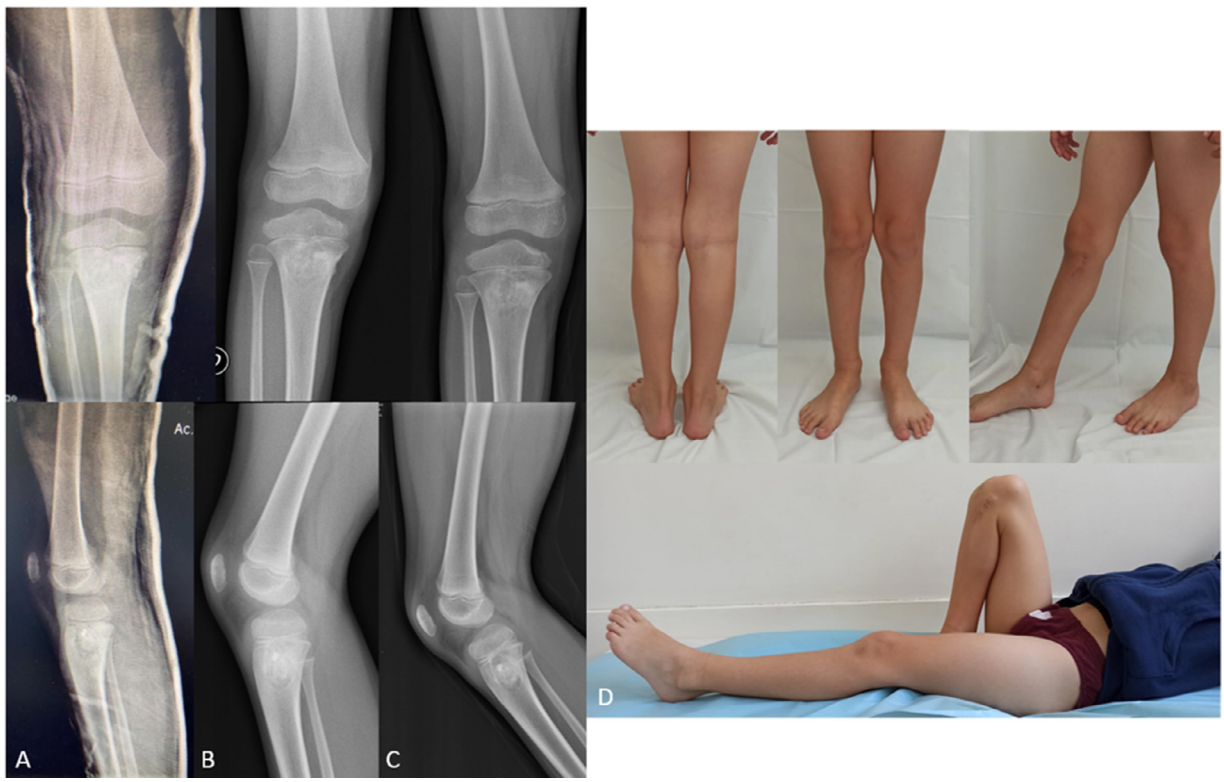

Fig. 7 – Postoperatory radiographic progress at 2 weeks (A), 2 months (B), and 6 months (C) from the definitive treatment. (D) Clinical images of the patient at 6-month follow-up demonstrating full range of motion of the right knee.

the presence of sheets of chondroblasts imbedded in an eosinophilic chondroid matrix and lace-like calcifications. Occasional giant cells and cellular atypia may be present [20]. Secondary aneurysmal bone cyst-type degeneration can be observed; however, this finding is more commonly noted in tumors of the hand and feet [3]. Prior studies have shown an elevated rate of misdiagnosis associated with this condition; the most frequent inaccurate diagnoses being giant cell tumor, aneurysmal bone cyst, and chondromyxoid fibroma; consequently, analysis by an experienced pathologist is paramount [21]. Additionally, in questionable or inconclusive cases staining with H3F3B or testing for the K36M mutation can be of help to elucidate the final diagnosis [22].

The treatment of chondroblastoma may vary depending on the location and extension of lesion, however, in most cases involves an intralesional curettage along an adjuvant,

such as phenol or hydrogen peroxide [2]. The void left inside the bone can then be filled with autograft, allograft or bone substitutes. Alternative treatment options include radiofrequency ablation for small lesions, and in the particular case of chondroblastomas within the proximal femur epiphysis, a trapdoor technique has been described [23,24]. Recurrences are noted to occur at a frequency of 10%-35% and, even though this tumor is considered to be a benign lesion, pulmonary metastases have been reported [25,26]. No histological feature was associated with an increased likelihood of recurrence or metastatic development [21]. Clinically, tumor location in the temporal bone or femoral head, as well as an incomplete resection without adjuvants, has been linked to a higher risk of recurrence [2,27].

The extraordinary location of the lesion in our patient as well as the presence of fluid-fluid levels in the MRI (Fig. 4), likely due to a secondary aneurysmal bone cyst degeneration of the chondroblastoma, led the treating team to propose additional differential diagnoses other than chondroblastoma. Furthermore, although chondroblastomas can present in a young population, the mean age of presentation averages between 11 and 13 years of age; our patient being quite younger obscured the initial diagnosis even further [28,29]. An initial biopsy properly performed allowed confirmation of the correct diagnosis and the appropriate treatment to be indicated. Experienced pathologists as well as a qualified multidisciplinary treatment team are paramount in such exceptional scenarios.

Conclusion

Chondroblastomas are benign tumors usually located in the epiphyses and apophyses of long bones of the immature skeleton. Moreover, these tumors typically present somewhat classical radiologic features. However, the presence of tumors outside the traditional location or epidemiological spectrum, along with the potential for histopathological misdiagnosis, can pose a challenge for the treating team.

Patient consent

Per the local institutional review board consent was exempt due to this being the case of research involving the collection or study of existing data, documents, records, pathological specimens, or diagnostic specimen with the information being recorded by the investigator in such a manner that subjects cannot be identified, directly or through identifiers linked to the subjects. Nevertheless, the patient and the legal guardian were informed and consented to publication.

REFERENCES

[1] Limaiem F, Tafti D, Rawla P. Chondroblastoma. StatPearls In. StatPearls Publishing; 2022.

[2] Deventer N, Deventer N, Gosheger G, de Vaal M, Budny T, Laufer A, et al. Chondroblastoma: is intralesional curettage with the use of adjuvants a sufficient way of therapy? J Bone Oncol 2020;26:100342. doi:10.1016/j.jbo.2020.100342.

[3] Chen W, DiFrancesco LM. Chondroblastoma: an update. Arch Pathol Lab Med 2017;141(6):867–71. doi:10.5858/arpa.2016-0281-RS.

[4] Alkadumi M, Duggal N, Kaur S, Dobtsis J. Chondroblastoma of the knee in a teenager. Radiol Case Rep 2021;16(12):3729–33. doi:10.1016/j.radcr.2021.08.065.

[5] Bloem JL, Mulder JD. Chondroblastoma: a clinical and radiological study of 104 cases. Skelet Radiol 1985;14(1):1–9. doi:10.1007/BF00361187.

[6] Papke DJ, Hung YP, Schaefer IM, Bredella MA, Charville GW, Reith JD, et al. Clinicopathologic characterization of malignant chondroblastoma: a neoplasm with locally aggressive behavior and metastatic potential that closely mimics chondroblastoma-like osteosarcoma. Mod Pathol 2020;33(11):2295–306. doi:10.1038/s41379-020-0604-2.

[7] Wu CT, Chen AC, Wang CJ, Wang CW, Fu CJ, Wong YC. Chondroblastoma of the triquetrum. Pediatr Neonatol 2013;54(4):278–80. doi:10.1016/j.pedneo.2012.11.017.

[8] Mu H, Jiang Y, Xue L, Hua Y, Lin J, Cai Z. H3.3 K36M mutation as a clinical diagnosis method of suspected chondroblastoma cases. Orthop Surg 2021;13(2):616–22. doi:10.1111/os.12878.

[9] Kolodny A. Bone sarcoma: the primary malignant tumors of bone and the giant cell tumor. Surg Gynec Obstet 1927;44(suppl 1):1–214.

[10] Codman EA. Epiphyseal chondromatous giant cell tumors of the upper end of the humerus. Surg Gynec Obstet 1931;52:543–8.

[11] Yang J, Tian W, Zhu X, Wang J. Chondroblastoma in the long bone diaphysis: a report of two cases with literature review. Chin J Cancer 2012;31(5):257–64. doi:10.5732/cjc.011.10402.

[12] Maheshwari AV, Jelinek JS, Song AJ, Nelson KJ, Murphey MD, Henshaw RM. Metaphyseal and diaphyseal chondroblastomas. Skelet Radiol 2011;40(12):1563–73. doi:10.1007/s00256-011-1227-y.

[13] Demertzis JL, Kyriakos M, Connolly S, McDonald DJ. Surface-based chondroblastoma of the tibia: a unique presentation. Skelet Radiol 2015;44(7):1045–50. doi:10.1007/s00256-015-2142-4.

[14] Sauer MA, Stegelmeier P, Crim JR, Layfield LJ, Evenski A. A report of an intracortical chondroblastoma of the diaphysis in a skeletally mature patient. Skelet Radiol 2022;51(12):2341–6. doi:10.1007/s00256-022-04088-4.

[15] Blancas C, Llauger J, Palmer J, Valverde S, Bagué S. Manifestaciones radiológicas del condroblastoma [Imaging findings in chondroblastoma]. Radiologia 2008;50(5):416–23. doi:10.1016/s0033-8338(08)76057-0.

[16] Punit A, Nadkarni S, Doomra T. Chondroblastoma of diaphysis of radius in a seven-year-old child. J Orthop Case Rep 2014;4(3):32–5. doi:10.13107/jocr.2250-0685.191.

[17] Choi J, Darrow MA, Zeitlinger L, Thorpe SW, Bindra J. Unusual chondroblastoma of the hand with large extraosseous soft tissue component. Radiol Case Rep 2022;17(9):3214–18. doi:10.1016/j.radcr.2022.06.024.

[18] Lambert J, Verstraeten T, Mermuys K. Chondroblastoma: an unusual cause of shoulder pain in adolescence. J Belg Soc Radiol 2016;100(1):16. doi:10.5334/jbr-btr.1027.

[19] Wang F, Li J, Yu D, Wang Q. Chondroblastoma of the distal femoral metaphysis: a case report with emphasis on imaging findings and differential diagnosis. Medicine 2018;97(17):e0336. doi:10.1097/MD.0000000000010336.

[20] Kurt AM, Unni KK, Sim FH, McLeod RA. Chondroblastoma of bone. Hum Pathol 1989;20(10):965–76. doi:10.1016/0046-8177(89)90268-2.

[21] de Silva MV, Reid R. Chondroblastoma: varied histologic appearance, potential diagnostic pitfalls, and clinicopathologic features associated with local recurrence. Ann Diagn Pathol 2003;7(4):205–13. doi:10.1016/s1092-9134(03)00048-0.
[22] Amary MF, Berisha F, Mozela R, Gibbons R, Guttridge A, O'Donnell P, et al. The H3F3 K36M mutant antibody is a sensitive and specific marker for the diagnosis of chondroblastoma. Histopathology 2016;69(1):121–7. doi:10.1111/his.12945.
[23] Sulaiman SRK, Al-Zubaidi SAM, Sakrana AA. Radio frequency ablation for the treatment of appendicular skeleton chondroblastoma: is it an excellent alternative? Systematic review and meta-analysis. Indian J Radiol Imaging 2022;32(4):523–30. doi:10.1055/s-0042-1755248.
[24] Katagiri H, Takahashi M, Murata H, Wasa J, Miyagi M, Honda Y. Direct femoral head approach without surgical dislocation for femoral head chondroblastoma: a report of two cases. BMC Surg 2022;22(1):327. doi:10.1186/s12893-022-01766-x.
[25] Binesh F, Moghadam RN, Abrisham J. A fatal case of pure metaphyseal chondroblastoma. BMJ Case Rep 2013;2013 bcr2013010315. doi:10.1136/bcr-2013-010315.
[26] Wing C, Watal P, Epelman M, Infante J, Chandra T. Pulmonary metastases of chondroblastoma in a pediatric patient: a case report and review of literature. Cureus 2022;14(9):e28897. doi:10.7759/cureus.28897.
[27] Laitinen MK, Stevenson JD, Evans S, Abudu A, Sumathi V, Jeys LM, et al. Chondroblastoma in pelvis and extremities—a single centre study of 177 cases. J Bone Oncol 2019;17:100248. doi:10.1016/j.jbo.2019.100248.
[28] Huang C, Lü XM, Fu G, Yang Z. Chondroblastoma in the children treated with intralesional curettage and bone grafting: outcomes and risk factors for local recurrence. Orthop Surg 2021;13(7):2102–10. doi:10.1111/os.13153.
[29] El-Ali AM, Coblentz A, Degnan AJ. Solitary long-bone epiphyseal lesions in children: radiologic-pathological correlation and epidemiology. Pediatr Radiol 2020;50(12):1724–34. doi:10.1007/s00247-020-04752-8.